

IN THE SPECIFICATION:

Please amend and replace paragraph [0026] at page 7 as follows:

[0026] Figure 1A, 1B and 1C Figs. 1-3 shows the GLC-MS analysis of SLOS plasma. (A) Fig 1 shows the three peaks detected at 22.0, 22.5 and 23.8 min; (B) Fig. 2 indicates that the first peak has the ions characteristics of 27-hydroxycholesterol; and (C) Fig. 3 indicates the third peak has the ions characteristic of 27-hydroxy-7-dehydro- and 8-dehydrocholesterol.

Please amend and replace paragraph [0027] at page 7 as follows:

[0027] Figure 2A, 2B and 2C Figs. 4-6 is the GLC-MS analysis of cholesta-5,7-diene-3 β ,27-diol (27-hydroxy-7-dehydrocholesterol). (A) Fig. 4 depicts the retention time (23.8 min) of the standard, cholesta-5,7-diene-3 β ,27-diol prepared by a known method (20). The complete mass spectral pattern of the standard is shown in B Fig. 5, and the mass spectral pattern of the peak obtained from a pooled plasma sample from patients with SLOS is shown in (C) Fig. 6.

Please amend and replace paragraph [0028] at page 7 as follows:

[0028] Figure 3 Fig. 7 shows the correlation of SLOS patient plasma sterol and 27-hydroxysterol levels.

Please amend and replace the Abstract paragraph at page 35, lines 6-12, as follows:

The invention provides methods of reducing the cholesterol accumulation in a subject, including methods of reducing cholesterol synthesis and methods of increasing cholesterol degradation. In accordance with the methods of the invention, cholesterol synthesis is inhibited by administering a compound capable of increasing 27-hydroxy-7-dehydrocholesterol and/or 27-

hydroxy-8-dehydrocholesterol levels, wherein an increase in 27-hydroxy-7-dehydrocholesterol and/or 27-hydroxy-8-dehydrocholesterol levels results in an inhibition of cholesterol synthesis.

~~Cholesterol degradation is increased by increasing the level of 7 α -hydroxylase in extrahepatic tissue and cells.~~ The invention includes screening methods and assays for identifying agent compounds capable of inhibiting 27-hydroxy-7-dehydrocholesterol reductase activity and of increasing 27-hydroxylation and 27-hydroxy-7-dehydrocholesterol and/or 27-hydroxy-8-dehydrocholesterol levels.